JACC: CASE REPORTS VOL. 3, NO. 2, 2021
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MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

BEGINNER

CASE REPORT: CLINICAL CASE

A Rare Cause of Angina

A Single Coronary Artery in Congenitally Corrected Transposition of the Great Arteries

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ABSTRACT

We report a man with congenitally corrected transposition of the great arteries and moderate anteroseptal ischemia; and cardiac computed tomography showed a single coronary artery with origin from the right aortic sinus. The perfusion of the morphological right ventricle by a single right coronary artery may be a cause of ischemia. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2021;3:202–5) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 44-year-old man presented to the emergency department with 2 months of chest pain that radiated to the neck and left arm at rest. On examination, blood pressure was 132/85 mm Hg, heart rate was 85 beats/min, and heart and lung auscultation findings were normal. An electrocardiogram was performed and showed Q waves in the right pre-cordial leads, Q-wave in V_1, and nondeep negative T waves in leads V_2 to V_6. Serum cardiac biomarkers were in normal ranges. A chest radiography was performed showing mesocardia, a normal cardiac index, and an egg-shaped ventricle; the liver shadow and stomach bubble were located normally (**Figure 1**). Then, a transthoracic echocardiogram was performed on the day of admission, showing levocardia as well as atrioventricular and ventriculoarterial discordance, suggestive of congenitally corrected transposition of the great arteries (CCTGA). Ejection fraction (EF) of the morphological right ventricle (systemic

LEARNING OBJECTIVES

- To understand that coronary artery anomalies can occur in up to 45% of patients with CCTGA.
- To recognize the inadequate coronary arterial supply as a possible cause of myocardial perfusion defects in patients with CCTGA.
- To recognize noninvasive imaging techniques as a safe method to evaluate congenital heart disease.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received September 20, 2020; revised manuscript received October 19, 2020, accepted November 2, 2020.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2020.11.044

ventricle) was 45%, and the left ventricular EF was 42%. Therefore, the patient was discharged home on the day of admission.

MEDICAL HISTORY

The patient had a history of CCTGA, systemic arterial hypertension, and hypercholesterolemia. He underwent open repair of an atrial septal defect at age 14 years.

DIFFERENTIAL DIAGNOSIS

Because of the initial presentation and cardiovascular risk factors, the differential diagnosis should include atheromatous coronary vascular disease.

INVESTIGATION

Ambulatory nuclear stress myocardial perfusion imaging showed moderate anteroseptal ischemia. Cardiac computed tomography (CT) was then performed to assess coronary artery anatomy. CT angiography performed in a 64-multislice scanner showed that the aorta originated from the morphological right ventricle and the pulmonary artery from the morphological left ventricle. The aorta originated immediately in front of the pulmonary artery. The left atrium was connected to the heavily trabeculated morphological right ventricle. The right atrium was connected to a morphological left ventricle, from which the pulmonary artery originated (**Figure 2**). It also showed that a single coronary artery originating from the right aortic sinus gave origin to the left main coronary artery and right coronary artery (Video 1, **Figure 3**). The morphological right ventricle EF was 41.9%. Cardiac CT showed no coronary artery disease, with a coronary CT calcium score of 0 Agatston Units.

MANAGEMENT

Treatment with metoprolol 25 mg twice a day was initiated. Also, the patient received enalapril 10 mg twice a day.

FOLLOW-UP

Within 1 year of follow-up in the cardiology clinic, the patient was in New York Heart Association functional class I. Ambulatory echocardiography showed no changes in ventricular function.

DISCUSSION

CCTGA was first described by Von Rokitansky in 1875; it is a rare defect representing approximately 0.5% of all congenital heart diseases, with the incidence reported to be 1/33,000 (1). This condition is characterized by discordance between the atrioventricular and ventriculoarterial connections; it results from leftward looping (L-loop) of the primitive cardiac tube instead of the normal rightward looping (D-loop) (2). It has been shown to be associated with different anomalies such as ventricular septal defect, pulmonary stenosis, valvular abnormalities, and coronary anomalies (3). In terms of the coronary artery anatomy, most patients with CCTGA have coronary artery ventricular concordance, with the morphological left ventricle supplied by a branching vessel similar to the left anterior descending and circumflex vessels and the morphological right ventricle supplied by a vessel similar to a right coronary artery (4). We describe a rare case of a single coronary artery in a patient with CCTGA.

Previous reports have described many anatomic anomalies of the coronary arteries in patients with CCTGA because coronary anomalies can be associated with up to 45% of cases (5). There is most often a mirror-image distribution of the coronary arteries, which follow the corresponding ventricles. The epicardial distribution of the right-sided coronary artery is that of a morphologically left coronary artery (circumflex and anterior descending coronary

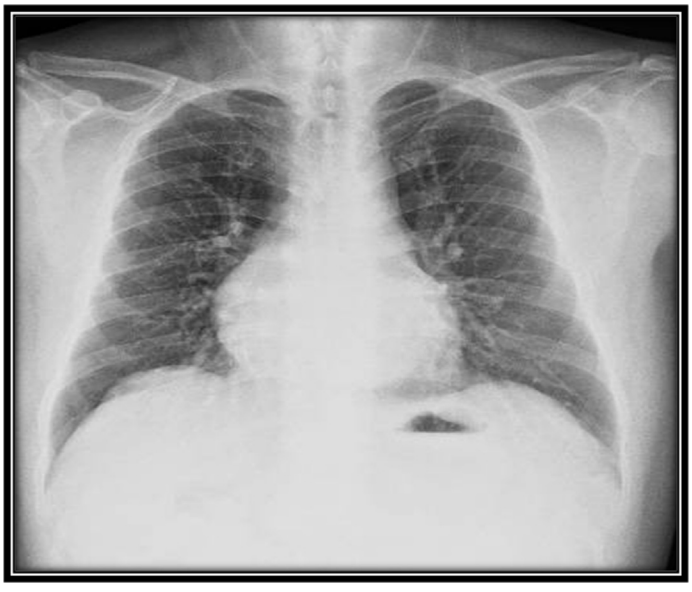
ABBREVIATIONS AND ACRONYMS

CCTGA = congenitally corrected transposition of the great arteries

CT = computed tomography

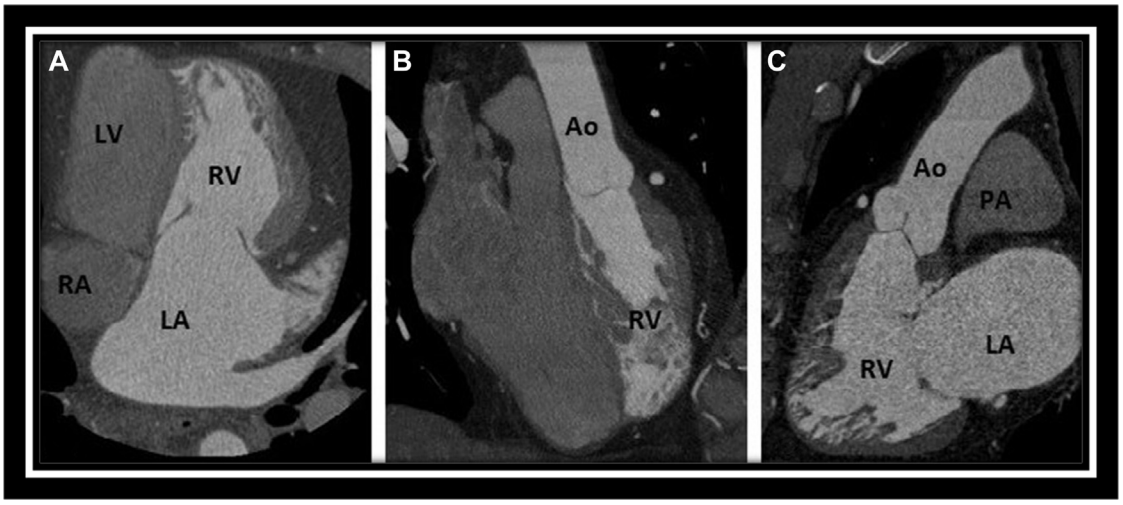
EF = ejection fraction

FIGURE 1 Chest Radiography



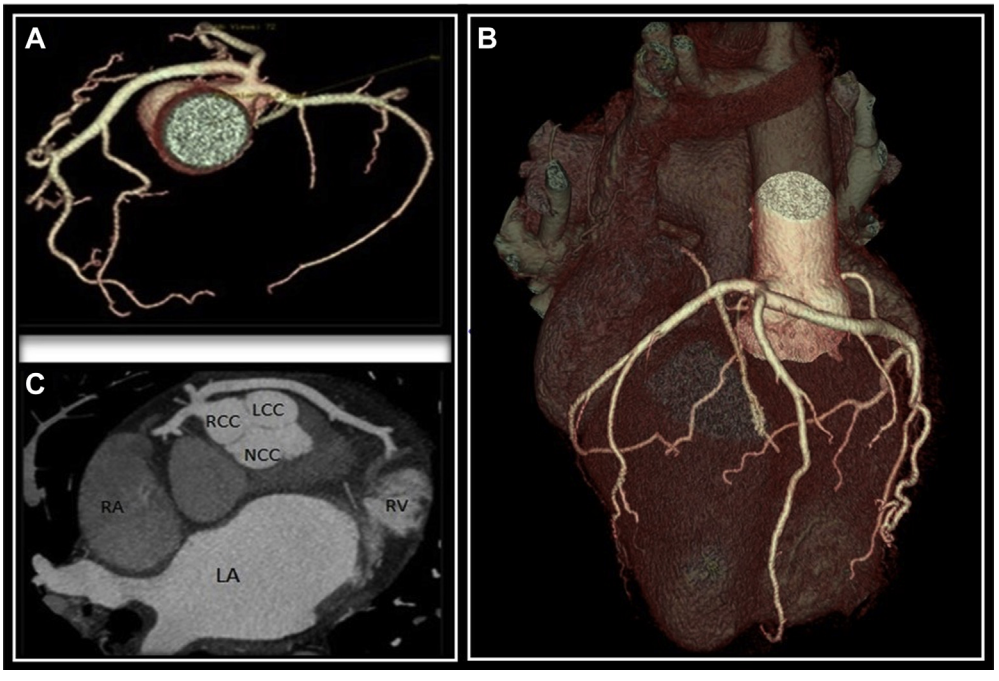
Chest x-ray film with cardiac contours classically described as appearing like an egg on string.

FIGURE 2 Computed Tomography Angiography



(A) Left atrium (LA) connected to the morphological right ventricle (RV). **(B)** Aorta originated from the morphological RV and the pulmonary artery (PA) from the left ventricle (LV). **(C)** Aorta origin located in front of the PA. Ao = aorta; RA = right atrium.

FIGURE 3 Single Coronary Artery



(A, B) Computed tomography 3-dimensional volume rendering technique reconstruction. A single coronary artery is observed. **(C)** A single coronary artery from the right coronary aortic sinus is observed. LCC = left coronary cuspid; NCC = noncoronary cuspid; RCC = right coronary cuspid; other abbreviations as in Figure 2.

arteries), and the left-sided coronary artery follows the left atrioventricular groove and supplies the right ventricle (4). In this case, the patient had just 1 ostia located in the right aortic sinus. The right ventricular coronary arterial supply may be inadequate for long-term ventricular function at systemic pressures, and that silent progressive irreversible ischemia of the right ventricular myocardium may make an important contribution to the development of right ventricular dysfunction and symptoms (6).

CONCLUSIONS

The newer noninvasive imaging techniques provide a safer method of investigating congenital anomalies and resulted in improved spatial orientation of the vessels and chambers of the heart. In this patient with CCTGA, no coronary artery disease was shown by cardiac CT. It was useful for elucidating the rare and unexpected congenital etiology of chest pain by the simultaneous evaluation of cardiac morphology and function as a single study, demonstrating the rare case of a single coronary artery. The morphological right ventricle that is perfused by a single right coronary artery may be the cause of reduced myocardial perfusion and the symptoms of this patient.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS angina, congenitally corrected transposition of the great arteries, single coronary artery

APPENDIX For a supplemental video, please see the online version of this paper.